494 Multiple brain abscesses in Cystic Fibrosis

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Aims: To describe a severe complication of cystic fibrosis involving cerebral abscess formation.
Methods: An 18 year old male diagnosed with cystic fibrosis at age 8 months presented with a 3 week history of fever, cough, and sputum. He was known to be colonized with *Klebsiella pneumoniae* and *Pseudomonas aeruginosa* (mucoid strain) and was chronically on nebulized tobramycin and oral ciprofloxacin. The day of admission he rapidly developed bilateral occipital headache, neck pain and stiffness, as well as nausea with vomiting.
Results: The admission WBC was 29K. His chest radiograph showed very severe bronchiectasis essentially destroying the right lung and sputum culture was positive for *P. aeruginosa* (mucoid strain). The CT head revealed multiple intracranial abcesses and left lateral ventriculits. A lumbar puncture revealed purulent fluid but culture results were all negative. Despite aggressive antibiotic and neurosurgical interventions, the patient deteriorated and died.
Discussion: Brain abscesses have been rarely reported in patients with cystic fibrosis with fewer than 15 reported cases in the English language literature. Predisposing factors appear to be sinusitis and immunosuppression. Previous reports have found pseudomonas, streptococcus, blastomycosis, and mouth organisms as the infectious etiology. In addition, no specific organism was found in some cases. There have been only two previous reports of multiple brain abscesses. In only one previous report did the patient survive the brain abscess. It is important to consider brain abscess in febrile cystic fibrosis patients who have headache or meningeal symptoms and start antibiotic therapy promptly.

495 Kidney pathology in children with Cystic Fibrosis

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Aim: To determine the character of kidney disease in children with cystic fibrosis (CF), elaborate its diagnostic criteria and treatment.
Methods: Included renal ultrasonography, measurements of the urinary concentration of Na+, K+, Ca2+, oxalate, phosphate, urates, creatinine, carbohydrates, urinary enzymes. 55 CF patients (1.5 to 16 years old) with severe and moderate disease were examined. There was a comparison group; 37 children with connective tissue disease (not CF) and a control group; 15 healthy children.
Results: Pathologies characterized as metabolic nephropathy were disclosed in 80% of CF subjects. Urinalysis showed various transitory pathologies (including protein-, leukocyte-, and hematuria) in 40% of CF children. The increase of occurrence of hyperoxaluria and calciuria was reliably determined in the older patients. Ultrasonography data indicated that in patients >5 yrs the increase of the echogenicity of kidney parenchyma and reduction of layer differenciation occurred more often. Uraturia and calcium-oxaluria, elevated excretion of carbohydrates and ultrasound evidences of nephropaty occurred more often in the subjects with severe CF than in those with moderate one. Irrespective of the age CF children showed the elevated activity of urine enzymes: lactatdehydrohenase, alkaline phosphatase and gammaglutamyltransferase. These results suggested the localization of the pathology in the renal proximal tubules and Henle loop. Metabolic nephropathy was successfully treated by membrane-stabilizing diphosphonate "Ksidifon", 15 mg/kg/day, administrated in the second half of a day, for 1 month.
Conclusion: We conclude that metabolic nephropathy is a widespread disease in CF subjects. The probable reasons – long-term therapy with nephrotoxic drugs and hypoxia leading to the cell membrane destabilization.

496 A longitudinal study of bone mineral density change in adults with Cystic Fibrosis

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Aims: Significant declines in bone mineral density (BMD) have been reported in adults with cystic fibrosis (CF) but longitudinal data greater than 1 year are scarce. The aim of this study was to document changes in BMD over a 3–4 year period and to investigate associated factors.
Methods: Data were collected from dual energy x-ray absorptiometry (DXA) scans in 86 CF adults (48 male, mean age 35.9). Absolute BMD (g/cm^2) of the total hip (TH), femoral neck (FN) and lumbar spine (LS) were recorded at 2 time points, 3–4 years apart. FEV1, Body Mass Index (BMI), number of intravenous (IV) antibiotics, steroid use, serum 25 OHD and testosterone levels were also recorded.
Results: BMD dropped significantly over 4 years at the FN ($p = 0.016$), but not at the TH or LS. The median % change was −5.28% (IQR −10.15—1.84) at the FN, −2.45% (IQR −6.97–0.41) at the TH, and −1.03% (IQR −5.67–1.90) at the LS. % Change BMD at the FN correlated with steroid use ($p = 0.02$). % Change BMD at the TH correlated with FEV1 ($p = 0.01$), BMI ($p = 0.003$), IV antibiotic use ($p = 0.02$) and steroid use ($p = 0.008$). % Change BMD at the LS correlated with FEV1 ($p = 0.01$), BMI ($p < 0.001$) and steroid use ($p < 0.001$). 21/86 (24.42%) lost BMD at 3 sites, 11/86 (12.79%) lost at 2, 24/86 (27.91%) lost at 1 and 30/86 (34.88%) remained stable or gained bone at all 3 sites.
Conclusion: Accelerated bone loss occurs to a greater degree at the femoral neck and is significantly related to steroid use in adults with CF. While bone loss occurs in many patients, some remain stable or gain BMD. This should be considered before commencing bisphosphonate therapy and supports the use of serial scans to document bone loss.

497 The determinants of daily physical activity (in steps/day) in Cystic Fibrosis patients: an observational study

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Study Objective: To examine the determinants of daily physical activity in patients with Cystic Fibrosis (CF)
Methods: Nineteen adults (13/6 m/f, age 30.2±7.0 (S.D) yrs) with CF were enrolled in an observational cross-sectional study. A pedometer (Digiwalker SW-200) was used for four weeks by the subjects to assess daily physical activity. Pulmonary function (FEV1;VC), lean weight, and BMI were measured. Dyspnea was assessed with the MRC dyspnea scale, health-related quality of life with the CF Questionnaire (CFQ 14+), exercise capacity with the 12-level Shuttle Walk Test (SWT) and lower-body strength with the Chair Stand Test (CST).
Results: VC and FEV1 were 75±17% and 47±19% of predicted. BMI averaged 21.5±3.1. The subjects walked 728±280 m. in the SWT (maximum of the test 1020 m). Pedometer measurements showed an average number of 7409 (±3548) steps/day. This mean number of steps is not different from values found in healthy subjects. There was a large inter-individual, but a small intra-individual variation in steps. There was also some variation in steps made related to daily outdoor temperature. Exercise capacity (SWT), dyspnea and FEV1 were (significantly) the most relevant predictors of steps measured by pedometer.
Conclusions: Mean daily physical activity status measured as steps/day with a pedometer in CF patients does not differ from healthy subjects. Exercise capacity, FEV1, and dyspnea score are determinants of physical activity (steps/day) in CF patients.